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Surgical Management of the Infant With Coarctation of the Aorta and Ventricular Septal Defect

JEANNY K. PARK, MD,* RALPH B. DELL, MD, KENT ELLIS, MD,
WELTON M. GERSONY, MD, FACC
New York, New York

Clinical and cardiac catheterization data were collected from 39 infants with coarctation of the aorta and ventricular septal defect, 31 of whom were initially managed only by surgical repair of coarctation. Data were analyzed to determine mortality, morbidity, outcome and factors that might predict survival or the need for septal defect closure.

Of the eight patients who did not require surgical treatment before 3 months of age, seven underwent coarctation repair alone at a mean age of 2.3 years. Of the 23 infants managed with coarctation repair alone, before age 3 months, 9 needed no additional surgical treatment and 6 required early and 8 required late repair of the ventricular septal defect. Seven infants underwent coarctation repair and simultaneous pulmonary artery banding and one eventually required debanding after spontaneous closure of the septal defect. The overall mortality rate in this series was 10.3% (mean follow-up time 5.7 years). Of 39 infants, 16 (41%) never required a second operation for ventricular septal defect closure.

For patients who had only coarctation or coarctation repair with pulmonary artery banding at <3 months of age, ventricular septal defect size was categorized as small (<0.5 cm/m^2), moderate (<1 cm/m^2) or large (>1 cm/m^2) on the basis of defect size at operative repair or echocardiographic or angiographic assessment. Defect size did not necessarily correlate with the need for operative repair. Stepwise multiple regression analysis revealed that increased right to left ventricular peak systolic pressure ($p = 0.004$) and decreased systemic venous oxygen content ($p = 0.028$) were significantly predictive of the eventual need for ventricular septal defect repair.

Thus, most infants with coarctation of the aorta and ventricular septal defect do not require pulmonary artery banding or open heart closure of the ventricular septal defect at the time of coarctation repair. A significant number of infants will not require a second operation.

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Infants with coarctation of the aorta and an associated large ventricular septal defect experience comparatively higher morbidity and mortality rates than do those with coarctation alone (1–3). Controversy exists as to the optimal management of these patients. For infants who present with congestive heart failure, surgical intervention is usually necessary to relieve the coarctation. However, questions remain as to appropriate management of the ventricular septal defect. Total repair of both defects has been accomplished in a few cases (4,5), but such aggressive management is most often not required. Some surgeons (6–8) perform pulmonary artery banding, either concomitantly with coarctation repair or shortly thereafter, to control pulmonary overcirculation. After a period of growth, the patient undergoes debanding and closure of the ventricular septal defect. However, it has been documented (5,9,10) that the septal defect can become smaller or even close with time. Therefore, repair of the coarctation without pulmonary artery banding also has been advocated (5,11) to avoid a second operation in some patients.

The purpose of the present study was to evaluate the mortality, morbidity and outcome in 39 infants who underwent surgical coarctation repair alone as the predominant method of initial management. Clinical and cardiac catheterization data were examined in an attempt to determine which factors predict survival or the need for ventricular septal defect closure.

Methods

Study patients. The records were reviewed of 39 infants <3 months of age with coarctation of the aorta and ventricular septal defect who presented to Columbia-Presbyterian Medical Center with congestive heart failure between May 1962 and June 1989. All available inpatient and outpatient records, cardiac catheterization data, angiograms and operative and autopsy reports were analyzed.

Before operation, patients were managed with digoxin and diuretic drugs; several infants required treatment with intravenous dopamine and prostaglandin. Since 1971, most infants have undergone immediate repair of the coarctation

From the Division of Pediatric Cardiology, Departments of Pediatrics and Radiology, College of Physicians and Surgeons of Columbia University and the Presbyterian Hospital, New York, New York.

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*Present address and address for correspondence: Jeanny K. Park, MD, Oregon Health Sciences University, 3181 Southwest Sam Jackson Park Road, Portland, Oregon 97201.

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when necessary, followed by medical management of subsequent congestive heart failure. If congestive heart failure proved intractable, the patient underwent surgical closure of the ventricular septal defect.

Surgical procedures. Coarctation repair was accomplished by resection of the coarcted segment with end to end anastomosis of the aorta in 30 patients and by turndown of the left subclavian artery in 8. One patient has not required surgical intervention. Seven infants underwent pulmonary artery banding at the time of coarctation repair and one patient had closure of the ventricular septal defect only. The mean follow-up time is 5.7 years (range 0.2 to 17).

Treatment groups. For purposes of analysis, the patients were classified into four groups. Group I consisted of eight patients who underwent surgical intervention at ≥3 months of age (mean age at initial operation 2.3 years). Groups II to IV consisted of 31 patients who underwent operation at <3 months of age. Infants in group II underwent coarctation repair only (23 patients, mean age 1.6 months); those in group III had both coarctation repair and pulmonary artery banding (7 patients, mean age 1 month). The single patient in group IV had ventricular septal defect repair only.

Available hemodynamic and angiographic data from the initial cardiac catheterization procedure were reviewed. Classification of ventricular septal defect size was based on 1) measured size at operation if the defect was repaired within the 1st 3 months of life; 2) assessment of size by angiographic report; or 3) cardiac ultrasound. On this basis, infants in groups II and III were categorized as having a small (<0.5 cm/m^2), moderate (0.5 to 1 cm/m^2) or large (>1 cm/m^2) ventricular septal defect (12).

Statistics. Analysis of variance was utilized to compare hemodynamic data among groups I, II and III (the one patient in group IV was not included in this analysis). Stepwise multiple regression analysis was used to determine the correlation between death or the need for ventricular septal defect closure and various hemodynamic and clinical variables. The survival curve was estimated by use of the Kaplan-Meier product limit method.

Results

Patient profile (Table 1). Of the 39 infants with coarctation of the aorta and ventricular septal defect, 21 were male and 18 female. Among these patients, 85% presented at ≤4 weeks of age. Ventricular septal defect location was membranous in 26, muscular in 6, multiple in 3 and indeterminate in 4. Most patients had pulmonary hypertension; 29 of 30 patients had a peak systolic pulmonary artery pressure >30 mm Hg and 17 of 20 had a pulmonary vascular resistance >2 U/m^2.

Outcome (Fig. 1)

Group I (no operation before 3 months of age). All patients in group I were managed medically with digoxin and diuretic drugs. Seven of the eight patients eventually underwent coarctation repair at a mean age of 2.3 years (range 0.6 to 15) and six never required ventricular septal defect closure. In one patient who initially presented at 2 weeks of age with a left to right shunt >3:1, pulmonary hypertension and a 30 mm Hg coarctation gradient, the ventricular septal defect closed spontaneously and the coarctation exhibited a minimal gradient (follow-up time 14 years). In another patient, surgical repair of the coarctation was recommended in the 1st month of life, but was refused by the parents until the child was 7 months old. The child then underwent ventricular septal defect closure at 9 months of age. This patient subsequently died of pulmonary hypertension at age 6 years.

Table 1. Clinical Profile of 39 Patients

	Mean ± SD	Range
Age (weeks)	3 ± 2.8	0 to 12
Weight (kg)	3.6 ± 1.4	2 to 10.3
Q_p/Q_s (n = 30)	3.4 ± 2.1	1 to 8.3
Systolic PAP (mm Hg) (n = 32)	51 ± 16	14 to 85
PVR (U/m^2) (n = 23)	5.7 ± 4.2	1.1 to 18
LAP (mm Hg) (n = 23)	11 ± 5	5 to 22
LVEDP (mm Hg) (n = 26)	13 ± 5	6 to 20

LAP = mean left atrial pressure; LVEDP = left ventricular end-diastolic pressure; PAP = pulmonary artery pressure; PVR = pulmonary vascular resistance; Q_p/Q_s = pulmonary to systemic flow ratio.

Group II (initial operation: coarctation repair only). Among the 23 patients in group II, 9 have not required closure of the ventricular septal defect (mean follow-up time 4 years) since coarctation repair. The septal defect was small in three patients, moderate-sized in three and large in three. Over time, repeat echocardiography or cardiac catheterization, or both, has demonstrated that all of these ventricular septal defects either have spontaneously closed or have become hemodynamically insignificant.

Six patients required ventricular septal defect closure within 3 months of the initial coarctation repair (mean age at defect closure 1.9 months); two of the six died. Two had a moderate-sized and the remaining four had a large ventricular septal defect. One patient with a large ventricular septal defect died when complete heart block developed after simultaneous coarctation and septal defect repair were performed. The clinical course of the remaining five patients was characterized by the need for a continued stay after operation (>2 weeks) to manage severe congestive heart failure (that is, dependence on a respirator or failure to thrive despite use of digoxin and diuretic drugs). One patient who was classified as having a moderate-sized ventricular septal defect died with low cardiac output after repair of this defect.

The remaining eight patients underwent "late" ventricular septal defect repair (mean age at repair 28 months); one of these died. Five infants were classified as having a large ventricular septal defect. Four infants (all with a large defect) required a prolonged hospital stay similar to that seen

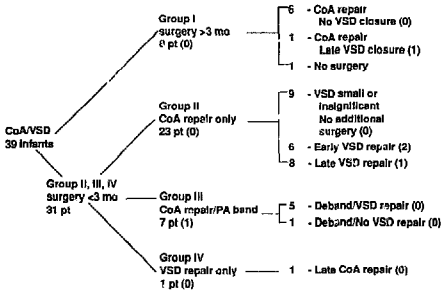

Figure 1. Outcome in 39 infants presenting with coarctation of the aorta (CoA) and ventricular septal defect (VSD) managed with or without pulmonary artery banding (PA band). Values in parentheses represent the number of deaths in patients with each intervention. pt = patient.

with the patients requiring early ventricular septal defect repair. However, a second operation was successfully postponed in these patients. The other four patients did well after operation and were discharged on treatment with digoxin and diuretic drugs. One patient died after undergoing late repair of multiple ventricular septal defects.

Group III (initial operation: coarctation repair and pulmonary artery banding). All seven infants in this group were categorized as having a large ventricular septal defect. Of these seven patients, one died shortly after operation. Five of the patients underwent subsequent debanding with ventricular septal defect closure at a mean of 18 months after coarctation repair and pulmonary artery banding. One patient who underwent debanding 6 years after the initial operation did not require ventricular septal defect closure.

Group IV (initial operation: ventricular septal defect repair only). The one infant in this group had a mild pressure gradient across the coarctation in the absence of a patent ductus arteriosus but required early ventricular septal defect repair because of congestive heart failure. The coarctation subsequently progressed, requiring repair 15 years later.

Survival

In the entire group of 39 patients, 4 died early and 1 died late (10.3% overall mortality rate). There have been no operative deaths since 1980. Figure 2 depicts the survival curve for these 39 patients, revealing that the greatest risk of death occurs in the 1st year of life, with a survival rate of 95% at 3 weeks, 92% at 13 weeks and 89% at 1.3 years. The survival rate then remains level until age 6 years, when it decreases to 85%. Sixteen patients have been followed up for >6 years (mean follow-up time 10.9 years), with no further increase in mortality rate.

Of the patients who have survived, further follow-up study revealed recurrent aortic coarctation in four, development of subaortic stenosis in four and valvular aortic stenosis in one patient; all nine patients required intervention. Mild mitral stenosis has been noted in two patients, although neither has undergone operative repair.

Predictors of the need for ventricular septal defect repair. Preoperative cardiac catheterization data did not differ among the treatment groups on a univariate basis (Table 2). Although there was no difference in the clinical variables that were predictive of death, stepwise multiple regression analysis indicated that an increased right to left ventricular peak systolic pressure ratio and a decreased systemic venous oxygen content were significantly predictive of the need for ventricular septal defect repair (Need for ventricular septal defect repair = 0.626 + [−0.103] [Systemic venous oxygen content] + 1.076 [Right to left ventricular peak systolic pressure ratio]).

Discussion

The management of infants with coarctation of the aorta and ventricular septal defect has been controversial (6–12).

Figure 2. Survival curve estimated by Kaplan-Meier product limit method for the 39 patients. Values in parentheses represent the number of patients who survived and were not lost to follow-up.

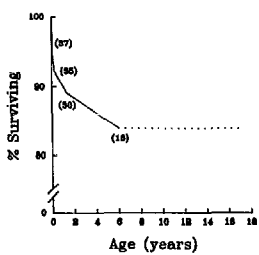

Table 2. Preoperative Cardiac Catheterization Data in 39 Patients

Catheterization Data	Group I (n = 8)	Group II (n = 23)	Group III (n = 7)
RV/LV peak systolic pressure	0.8 ± 0.2 (n = 5)	0.9 ± 0.2 (n = 17)	1 ± 0 (n = 5)
Pressures (mm Hg)			
PAP, systolic	66 ± 34 (n = 8)	83 ± 33 (n = 17)	91 ± 19 (n = 5)
PAP, diastolic	28 ± 13 (n = 8)	20 ± 10 (n = 17)	33 ± 9 (n = 5)
PAP, mean	45 ± 21 (n = 8)	52 ± 14 (n = 17)	58 ± 14 (n = 5)
LVEDP	12 ± 6 (n = 6)	13 ± 4 (n = 14)	12 ± 6 (n = 5)
LAP	9 ± 2 (n = 6)	11 ± 5 (n = 12)	12 ± 7 (n = 5)
Coarctation gradient	40 ± 25 (n = 7)	42 ± 21 (n = 15)	31 ± 17 (n = 5)
Systemic venous O_2 content (ml O_2/dl)	9.7 ± 1.7 (n = 8)	12.7 ± 1 (n = 17)	9.9 ± 1.7 (n = 5)
Q_P/Q_S ratio	2.8:1 ± 2.4 (n = 8)	3.4:1 ± 2.3 (n = 16)	4.3:1 ± 1 (n = 5)
Cardiac index (liters/min per m^2)	3 ± 0.8 (n = 8)	2.8 ± 0.8 (n = 15)	2.8 ± 0.7 (n = 5)
PVR (U/m^2)	6.3 ± 5.4 (n = 6)	6.3 ± 4.9 (n = 11)	4.2 ± 1.7 (n = 5)

There was no significant difference ($p < 0.05$) among mean values for all three groups. Numbers in parentheses indicate the number of patients with available catheterization data. O_2 = oxygen; RV/LV peak systolic pressure = right to left ventricular peak systolic pressure ratio; other abbreviations as in Table 1.

Performance of pulmonary artery banding before coarctation repair was first advocated in 1975 by Connors et al. (6) and further endorsed by other groups (3,7,8,13).

Other groups recommend against routine performance of pulmonary artery banding in this group of patients. Neches et al. (4) reported the outcome in 53 infants with aortic coarctation and ventricular septal defect. The ventricular septal defect closed spontaneously or became hemodynamically insignificant in 11 of 25 patients who were classified as having a large defect. Thus, these investigators (4) recommended that infants with aortic coarctation and ventricular septal defect should be managed with resection of the coarctation without simultaneous pulmonary artery banding. Other groups (1,2,11) have reported comparable results with similar conclusions.

Rationale for management. It has been reported (8) that the majority of infants with coarctation of the aorta and a ventricular septal defect have a small or normal left ventricular volume. This lack of volume loading may suggest why pulmonary artery banding or direct ventricular septal defect repair does not appear to be essential in initial surgical management. Adequate relief of increased left ventricular afterload is often sufficient to enable medical management of these patients (14).

Among infants who undergo pulmonary artery banding, a significant number have ventricular septal defects that close or become hemodynamically insignificant. Whether or not these infants eventually need ventricular septal defect repair, they always require pulmonary artery debanding, often with pulmonary angioplasty. In our series, one of the seven patients who underwent initial pulmonary artery banding subsequently required debanding without repair of the ventricular septal defect. Of the 23 infants who had coarctation repair only, 9 did not require a second operation for ventricular septal defect repair. If these infants had undergone pulmonary artery banding, there would have been nine unnecessary second operations.

Ventricular septal defect size. For patients who underwent coarctation repair only (group II), categorization of ventricular septal defect size as moderate or large did not necessarily correlate with the eventual need for early or late repair. Most patients who did not require a second operation to repair the ventricular septal defect had a small to moderate-sized defect. However, three patients with a large defect did not need surgical closure. A limitation of this study is that direct measurement of ventricular septal defect size by angiography or echocardiography, or both, was imprecise in patients who presented early in this long-term series.

Other attempts to discover which patients will benefit from early intervention of the ventricular septal defect (3,13) have included the use of stepwise multiple regression analysis. In our study this analysis revealed that high right to left ventricular peak systolic pressure ratio ($p = 0.004$) and low systemic venous oxygen content ($p = 0.028$) were predictive of the eventual need for surgical ventricular septal defect repair. In the absence of mitral or pulmonary venous obstruction, or both, an increased right to left ventricular peak systolic pressure ratio suggests the presence of a large nonrestrictive ventricular septal defect. A decreased systemic venous oxygen content implies decreased cardiac output and poor toleration of the added hemodynamic burden resulting from a large intraventricular communication.

Conclusions. Successful management of infants with aortic coarctation and ventricular septal defect who present with congestive heart failure is possible with repair of coarctation alone. As seen in this series, a large ventricular septal defect does not necessarily require immediate intervention and patients with moderate-sized defects are even more likely than those with larger defects to be able to avoid repeat surgery. Early ventricular septal defect repair or pulmonary artery banding should be considered in patients in whom medical management of postoperative congestive heart failure is unsuccessful. Statistical analysis suggests that the presence of both an unrestrictive ventricular septal defect and low venous oxygen content correlate with the need for ventricular septal defect repair.

At our institution, initial combined repair of both aortic coarctation and the ventricular septal defect would be undertaken, rather than pulmonary artery banding. Indications for pulmonary artery banding include 1) the presence of multiple ventricular septal defects; 2) a "huge" ventricular septal defect (>50% of the ventricular septum); or 3) com-

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plex congenital heart disease (that is, transposition of the great arteries with a ventricular septal defect or atrioventricular canal defect) (11). Nevertheless, in the majority of infants, aortic coarctation repair alone is sufficient, and in this series, 41% of the patients managed in this manner did not require another operation.

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